A Rare Case of a Pilar Cyst With Ductal Differentiation

Vanda F. Torous, MD, Albert Su, MD,† Scott W. Binder, MD,† and Seong H. Ra, MD†‡*

Abstract: Pilar cysts are common squamous-lined cysts that typically occur on the scalp. They are believed to arise from the isthmus of anagen hairs or from the sac surrounding catagen and telogen hairs. The authors describe a rare case of a pilar cyst with prominent ductal differentiation, presumably of eccrine derivation. Sweat duct differentiation has been described in a myriad of cutaneous neoplasms and rarely within epidermoid cysts. The authors could only find one other case in the literature describing a pilar cyst with sebaceous and apocrine differentiation. The clinicopathologic findings are described here.

Key Words: pilar cyst, trichilemmal cyst, ductal differentiation

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INTRODUCTION

Pilar cysts, also known as trichilemmal cysts, are common, typically solitary, dermal and/or subcutaneous cysts that preferentially occur in areas with dense hair follicle concentrations. They are the most common cutaneous cyst in the scalp[1,2] and the second most common in the head and neck. Pilar cysts occur in 5%–10% of the population with a strong female preponderance. Histologically, they show a 3-to-4-layer thick lining of keratinocytes with abrupt keratinization into compact keratin without a granular cell layer.[2] Ductal differentiation in a pilar cyst is exceptional, and to our knowledge, there has been only 1 case reported.[3] The second case of a pilar cyst with ductal differentiation is described here.

CASE REPORT

The patient is a 62-year-old woman who presented with a mobile 2 cm mass of the left parietal scalp. The clinical differential diagnosis included a cyst, and an excisional biopsy was subsequently performed. Histological sections showed a squamous-lined cyst with prominently interspersed ducts lined by flat epithelial cells (Figs. 1A, B). No apocrine snouts were identified. Immunohistochemical stains for epithelial membrane antigen and carcinoembryonic antigen polyclonal strongly highlighted the ducts (Figs. 1C, D). There was focal staining of ducts for S100. The ducts were negative for gross cystic disease fluid protein (GCDFP)–15. The cyst lining demonstrated an abrupt transition to homogenous keratin and no granular cell layer. The cyst cavity showed homogenous keratin with cholesterol clefts and occasional calcifications. A focal area of rupture, characterized by loss of the squamous lining, keratin granulomas, and granulation tissue, was seen. No dysplasia or malignancy was identified. The constellation of pathologic findings was consistent with a pilar cyst with ductal differentiation. Because of the focal S100 positivity of ducts and lack of apocrine snouts, an eccrine origin was favored. The cyst was completely excised at the time of surgery, and the patient is free of disease after 1 year.

DISCUSSION

Pilar cysts are thought to be derived from the external root sheath of the hair follicle and lined by stratified squamous epithelium similar to that in the isthmus of the hair follicle. In contrast to the typical pilar cyst, our case showed interspersed ducts that were believed to be eccrine. Although the presence of ductal differentiation within pilar cysts is rare, it is not surprising.[3] Divergent glandular and follicular differentiation has been described in association with myriad of cutaneous neoplasms including basal cell carcinoma,[4] trichilemmoma, inverted follicular keratosis, and mixed tumors.[5–8] Rare adnexal tumors have been shown to undergo trichogenic, apocrine, and eccrine differentiation.[5,6] It has been hypothesized that multidirectional differentiation of cutaneous neoplasms occurs through pluripotential cells of the epidermis and/or adnexal structures.[3,5,6] These findings are consistent with the theory of a common embryological origin for the 3 adnexal components as a follicular–sebaceous–apocrine unit and eccrine sweat glands from stratum germinatum.[5]

Based on this theory, the variety of adnexal combinations encountered within hybrid cysts is not surprising. The hybrid cyst refers to cysts with dual linings including variable combinations of epidermoid, trichilemmal, pilomatrical, steatocystoma, and eruptive vellus hair cyst features.[9,10] Also, hybrid cysts with follicular and apocrine differentiation have been described.[11,12] The more common epidermoid cysts have been described in association with eccrine ducts.[13]

There has only been one other report of a pilar cyst with glandular differentiation. Hanau and Grosshans[3] described a trichilemmal cyst with sebaceous and apocrine structures. It is surprising that more pilar cysts with divergent adnexal differentiation have not been described. It is possible that this phenomenon is more common and overlooked. Based on our observations, it would not be surprising if cases of pilar cysts with any combination of divergent follicular and/or glandular differentiation were encountered.

In conclusion, the description of this rare cyst has uncertain clinical significance and likely represents the presence of a pluripotential cell with the ability to undergo

From the *Department of Pathology and Laboratory Medicine, Beth Israel Deaconess Medical Center, Boston, MA; †Department of Pathology and Laboratory Medicine, David Geffen School of Medicine at UCLA, Los Angeles, CA; and ‡San Diego Pathology Medical Group, Inc, San Diego, CA.

The authors declare no conflicts of interest.

Reprints: Vanda F. Torous, MD, Department of Pathology and Laboratory Medicine, Harvard School of Medicine, 330 Brookline Avenue, Boston, MA 02215 (e-mail: vtorous@bidmc.harvard.edu).

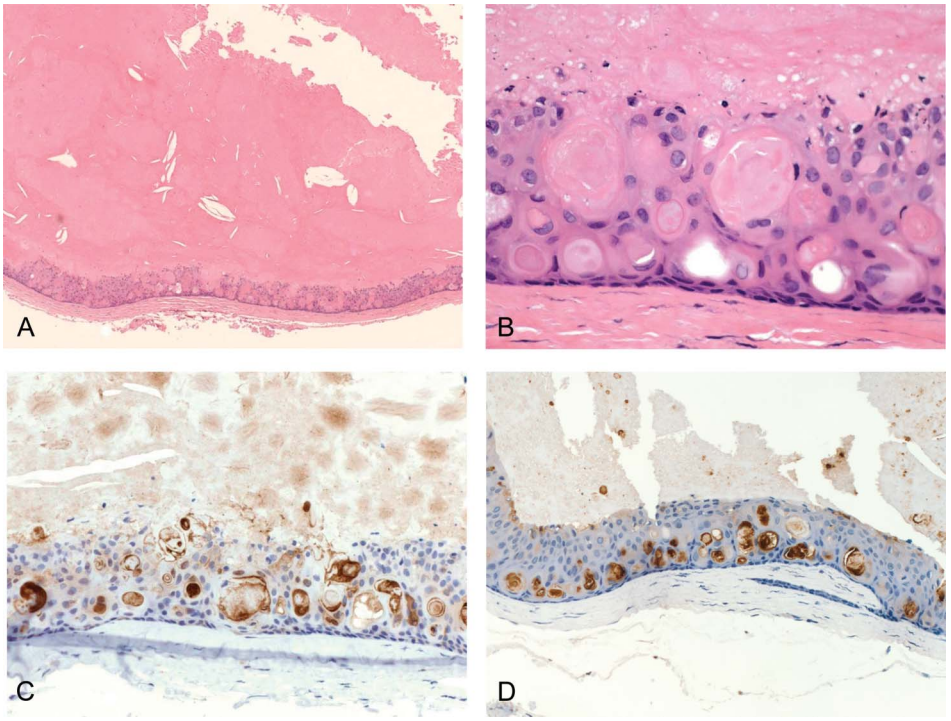

FIGURE 1. A, Microscopic examination of the cyst shows squamous lining with prominently interspersed ducts lined by flat epithelial cells. The cyst cavity demonstrates homogenous keratin with cholesterol clefts (H&E ×5). B, The cyst cavity shows an abrupt transition to homogenous keratin and no granular cell layer (H&E ×40). C, Immunohistochemistry stain for epithelial membrane antigen strongly highlights the ducts. D, Immunohistochemistry stain for carcinoembryonic antigen also strongly highlights the ducts.

divergent trichilemmal and eccrine ductal differentiation. As more cases of divergent differentiation within a pilar cyst are reported, it may be possible to ascertain whether the finding of ductal differentiation is merely a histological curiosity or has some prognostic significance.

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